CASE REPORT

Aneurysmal Bone Cyst Involving the Skull Base: Report of Three Cases

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ABSTRACT—Aneurysmal bone cyst is a benign, non-neoplastic lesion that present most frequently under the age of 20 years. The metaphysis of long bones is the usual site of origin. The involvement of the skull is rare. In the 2.5 to 6% of such cases reported in the literature, the skull vault is more often the site than the skull base. Three cases of aneurysmal bone cyst involving the skull base have been managed at King Faisal Specialist Hospital and Research Center. Two females and one male, all 10 years of age and younger, presented with a painless, progressive swelling. The preoperative radiological studies showed characteristic findings and were highly suggestive of the diagnosis. Angiography also gave characteristic findings. Preoperative endovascular embolization of the arterial feeders to the tumor was performed in two patients who had a significant decrease in intraoperative bleeding from the tumor. All cases underwent surgical excision with a good outcome.

CASE REPORTS

Case 1

A 6-year-old Saudi boy was noticed by his family to have a swelling on the left side of his face for 4 months prior to presenting at the hospital. A history of trauma was denied. The lesion was occasionally painful but not cumbersome. The child was admitted to the referring hospital where he underwent temporal craniotomy and attempted excision of the tumor. Physical examination on admission to this Institution revealed a hard swelling at the temporal region in front of the left ear. The lesion was not tender and the overlying skin was normal. No bruit could be heard on or surrounding the swelling. The facial nerve and all other neurological functions were intact. A plain X ray of the skull showed the site of the previous craniotomy. A destructive lesion was shown extending medially to the foramen ovale, posteriorly to the mastoid process, and anteriorly to the sphenoid wing of the temporal bone. Computed tomographic (CT) scan scanning showed a rounded bony lesion arising from the middle cranial fossa. The lesion extended mainly intracranially, compressing the temporal lobe and distorting and expanding the posterior part of the zygomatic process (Fig. 1). Fine trabiculae were seen within the lesion. The posterior part of the lesion enhanced intensely after contrast injection. Multiple fluid–fluid levels were seen. Magnetic resonance imaging (MRI) showed low signal intensity lesion on T1-weighted imaging extending into the base of the left temporal fossa and elevating the left temporal lobe (Fig. 2). The lesion extended into but did not involve the left temporo-mandibular joint. On T2-weighted imaging, the lesion showed areas of increased signal intensity consistent with subacute bleed; the lesion was loculated and contained multiple fluid–fluid levels. The patient was evaluated by neuro- and the maxillofacial surgeons and underwent total excision of the tumor piecemeal. This was performed in two stages; first, by the neurosurgeons for the intracranial expansion; and later, by the maxillofacial surgeons for the extension into the zygomatic process. At surgery the margins of the tumor were bony hard while the center was soft and compressible. Internally, the tumor was multilocu-

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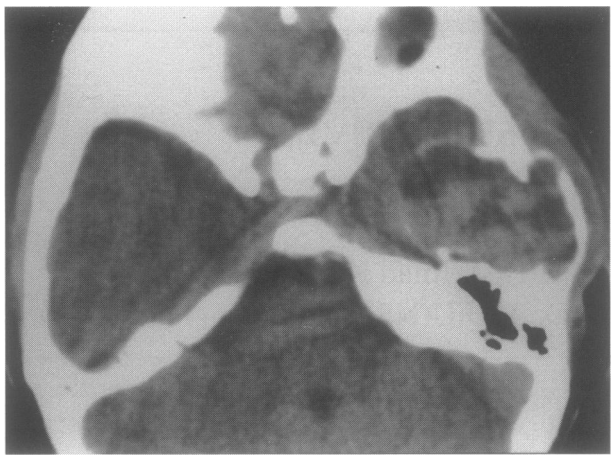

Figure 1. Noncontrasted CT scan of the head showing a temporal fossa lesion. The lesion is of multiple densities. Fluid levels are demonstrated between the lower hyperdense and the upper hypodense fluids.

lated and contained dark venous blood. Adhesion to the dura was noted but no intradural extension was present. The child tolerated both surgeries well with no complications. Histologic examination showed fragments of bony tissue separated by loose connective tissue containing dilated spaces. These spaces were filled with blood and surrounded by stroma-containing spindle cells and multinucleated giant cells; Benign reactive bone was present around the spaces. The diagnosis of aneurysmal bone cyst was made. The child was well with no recurrence 52 months postsurgery.

Case 2

This was a 1-year-old Saudi boy, the product of normal pregnancy and spontaneous vaginal delivery. The neonatal period and early infancy were unremarkable. During play the family noticed that the child did not see well or follow objects. He was evaluated by an ophthalmologist who declared him to be completely blind. There was no history of head trauma. The child was noticed to be a mouth breather and he had a horizontal nystagmus. Examination of other neurological functions and other systems was unremarkable. Complete blood count, renal and coagulation profiles all showed normal levels. A CT scan of the head revealed a large multicompartment lesion consisting of areas of different density. The tumor extended intracranially but extradurally from the mid-frontal base with erosion into the orbits bilaterally and the nasopharynx. Both T1- and T2-weighted MRI showed multiple fluid–fluid levels. Cerebral angiography showed tumor blush supplied by both internal maxillary arteries. Endovascular embolization of the feeders using micro-emboli resulted in 90% devascularization of the tumor. The intracranial portion of the tumor was exposed through a bifrontal craniotomy and the dura was dissected from the midline frontal skull base. A bony defect was evident at the area of the crista galae and a bony bulge was seen in the midline frontal base. The inner portion of the tumor was soft and could be scooped out with minimal bleeding. The otolaryngeologist performed a transnasal excision of the portion extending through the olfactory groove to the nasal cavity. The histologic examination revealed hemorrhagic lacunes surrounded by proliferating spindle cells. Giant cells were present and osteoid tissue was seen at the periphery of the lesion with prominent osteoblasts. Postoperatively, the child developed subgaleal cerebrospinal fluid collection, which disappeared gradually over weeks. A CT scan of the head showed areas of suspected recurrence but this was ruled out by biopsy. The child was well 8 months after excision of the tumor.

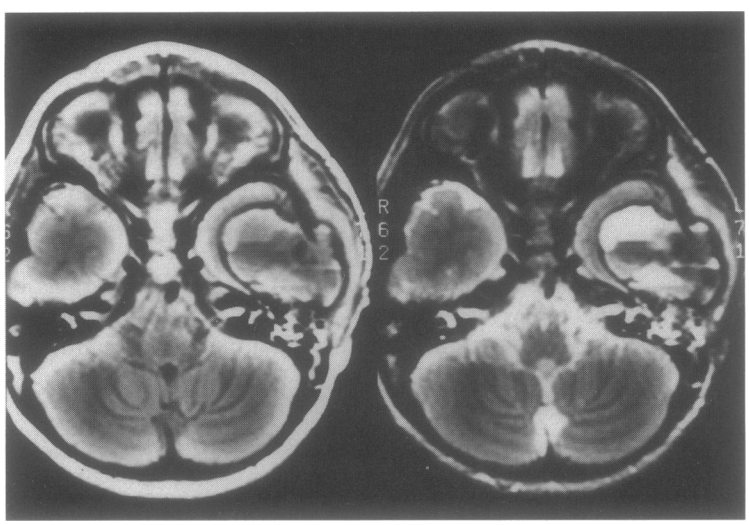

Figure 2. MRI of the head of the same patient in Figure 1. The fluid–fluid levels are clearly demonstrated.

Case 3

A 10-year-old Saudi girl presented with a bulge behind her left ear, which had first been noticed by her family 1 year earlier and it had progressively enlarged. The lesion was painless. Her family denied any head injury. Physical examination showed a hard mass measuring 4 ×5 cm behind the left ear, occupying the left lateral suboccipital region and extending to the left occipital bone superiorly. No other masses were noted. Examination of neurological function and other systems was unremarkable. Laboratory tests including complete blood count and renal and hematologic profiles, were normal. Plain X ray of the skull showed a large, rounded, intradeploic mass with calcified rim (Fig. 3) extending from the posterior mastoid area to the left occipital/suboccipital bone. The cortical bone was distorted inward and outward, but its consistency was normal. Faint calcification was evident within the lesion. A CT scan of the head showed an extra-dural mass with well-defined calcified margins causing significant mass effect. The content was unhomogeneous with calcified cystic and solid components. Some enhancement appeared in the solid portion after contrast injection. An MRI of the head revealed the cystic nature of the mass with fluid–fluid levels. The soft tissue component enhanced after gadolinium injection. Selective cerebral angiography showed tumor blush (Fig. 4). Branches of the external carotid artery fed the tumor. Catheterization of the feeders and embolization with *N*-butyl cyanoacrylate decreased the blush considerably. There was insignificant bleeding from the tumor at surgery. The bony bulge was deroofed. The content was soft, spongy, pinkish-colored material, which was removed in one piece. The inner table of the skull was bulging inward displacing and adherent to the dura but dissectable with no injury to the dura. The involved mastoid process was drilled out. Total removal was accomplished. Histologic examination was characteristic of aneurysmal bone cyst. At 8 months' follow-up, the child was completely well.

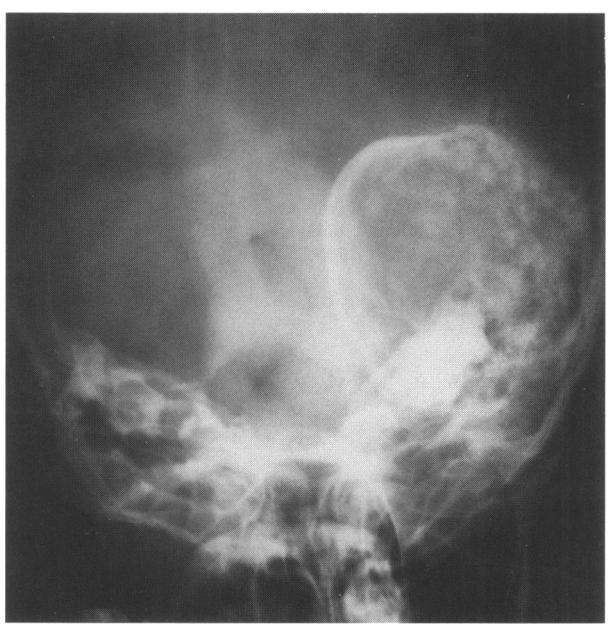

Figure 3. Plain X ray of the skull for the patient no. 3. Rounded tumor with a calcified rim is arising from the deploic space causing expansion. Fine trabeculation can be observed.

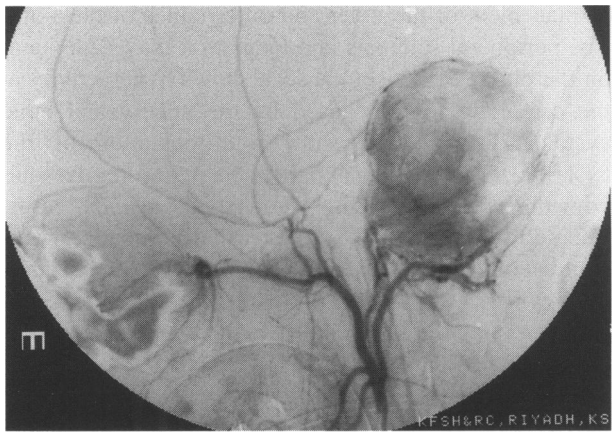

Figure 4. Selective cerebral angiography for the same patient in Figure 3. Intense tumor blush is demonstrated arising from branches of the external carotid artery.

DISCUSSION

Aneurysmal bone cyst is a benign, non-neoplastic lesion that present most frequently before the age of 20 years.[1–3] All our cases were aged 10 years or less. The metaphysis of the long bones is the usual site of origin.[2,3] The involvement of the skull is rarely reported,[4,5] and most of these appear on the vault.[6,7] All the cases in the present report originate from the skull base. Aneurysmal bone cyst was first described as a lesion with a "Blow-out" appearance of such lesions.[8]

Theories on the pathogenesis of aneurysmal bone cyst include the development of local hemodynamic alteration with secondary venous hypertension, as described by Lichtenstein.[3] Local trauma prior to the development of aneurysmal bone cyst has been reported.[5,7] Some bony lesions, such as fibrous dysplasia or chondroblastoma, have been demonstrated in associated with aneurysmal bone cyst.[4,9,10] These may have preceded the appearance of aneurysmal bone cyst and predisposed for its development.

The plain X ray picture of expansile " blow-out" or "soap-bubble" is characteristic.[9,11,12] The lesion originates within the deploic space causing expansion and

displacement of the inner and outer tables of the skull. The peripheral sclerosis and loculation may be evident on the plain X ray. Isotope scan shows hyperactivity at the osteolytic lesion caused by the aneurysmal bone cyst.[13] CT and MRI show multiloculation of the cyst.[4,14–16] The content is partly cystic and partly solid with the presence of fluid–fluid levels in most cases. The solid portion of the tumor usually enhances with contrast injection. Cerebral angiography often shows only evidence of mass effect with no pathological vessels supplying the lesion.[11,12,16] In the two cases with angiographic examination in this report, the tumor showed significant tumor blush. Endovascular embolization of the feeders was performed in both cases, which decreased the tumor blush on immediate postembolization angiography, and intraoperative bleeding was minimal in both cases. The use of endovascular embolization as a preoperative therapy has been mentioned in the literature for aneurysmal bone cyst involving the skull or other skeletal parts.[17,18]

Histologically, the lesion is characterized by the presence of multiple loculations, filled with venous blood and lined by spindle-shaped fibroblasts and with scattered multinucleated giant cells and stromal cells.[19,20]

The treatment option of choice for these lesions is total excision when possible, which is curative,[21,22] but difficult to accomplish in cases where the skull base is involved.[10,22] In these cases, partial resection and curettage of the inner contents may suffice to halt progression of the disease or even cause regression, but there may be recurrence. As these lesions are non-neoplastic and benign, the use of radiation therapy is not recommended, although it is reported in the literature. There is no role for chemotherapy.

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